

Incidental olfactory aplasia: a case report

We report a case of olfactory aplasia with no other cerebral malformation in an elderly woman of high intellect. The patient was 73 years old and first presented at the age of 69 complaining of forgetfulness. On review by a neurologist at this time, history and examination were unremarkable apart from information volunteered by the patient herself that she had never had any sense of smell.

Neuropsychological testing was also performed with standard tests from the Wechsler Intelligence scale. The patient achieved a memory quotient well in excess of 143, which is the highest score in the published norms. Her pro-rated verbal and performance IQs were also in the high range at 134.

She was admitted to the Alfred Hospital, Melbourne, in January 1992 after collapse. A cerebral CT scan showed an intracerebellar haemorrhage with extension of blood into the fourth, third, and lateral ventricles. She was taken to theatre where the blood clot was evacuated and a Rickham's reservoir was inserted. A postoperative cerebral digital subtraction angiogram revealed active bleeding from the left posterior inferior cerebellar artery. The patient then returned to theatre where a bleeding point was identified and trapped with two clips. She did not regain consciousness after the operation and showed no response to standard stimuli. She died two days later.

A postmortem of the brain only was performed. External examination showed normal facies and secondary sexual characteristics. An unexpected finding was that of absence of the olfactory bulbs and tracts (figure). This was associated with a deficiency of the falx cerebri anteriorly so that some of the medial frontal gyri of the right and left sides tended to interdigitate. The ventricular system was of normal configuration, however. There was extensive haemorrhagic necrosis of both cerebellar hemispheres, particularly the left, with upwards herniation bilaterally and mottled haemorrhagic necrosis of the mid-brain and pons. Review of the cerebral CT scan showed no detectable abnormalities of the skull.

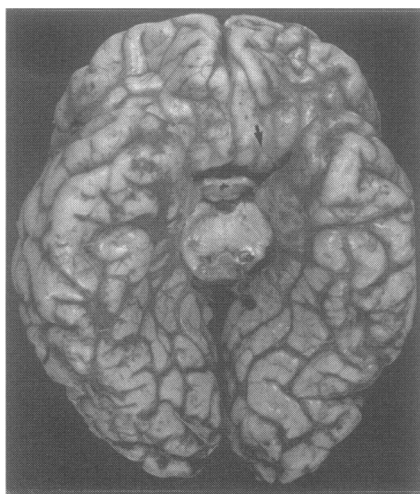


Figure 1 Base of brain showing absence of olfactory tracts, bulbs and olfactory sulci bilaterally. The olfactory tubercle is present (arrowed).

Histological examination of the frontal cortex and white matter was unremarkable. The pituitary gland appeared normal. Some hypoxic change was noted. There was extensive haemorrhagic necrosis of the mid-brain, pons, and the periventricular region of the medulla. Sections of the cerebellum showed a haemangioblastoma with focal necrosis and extensive haemorrhage.

Olfactory aplasia is rarely encountered as an isolated abnormality in an otherwise normal patient.¹ More commonly it is associated with a wide range of other malformations that may be both cerebral and somatic. The cerebral malformations are usually categorised by the broad term arhinencephaly, and include holoprosencephaly, agenesis of the corpus callosum, abnormalities of the optic tracts, cerebellar hypoplasia, and dentoolivary dysplasia.² These abnormalities may range from mild to severe in degree but mental retardation is usually present. Somatic malformations are also common. These include facial dysmorphism, which may take the classical form of midline hypoplasia, especially when associated with holoprosencephaly, with cyclopia, hypotelorism, ethmocephaly, cebocephaly, and cleft palate.^{3,4} A wide spectrum of other systemic abnormalities has been found, particularly when holoprosencephaly is associated with chromosomal anomalies,^{3,4} but also in other conditions where olfactory aplasia occurs as the sole macroscopic cerebral abnormality, such as Kallmann's Syndrome. This syndrome is characterised by hypogonadic hypogonadism and agenesis of the olfactory bulbs, which may be associated with diabetes mellitus, cleft lip and palate, sensorineural deafness, and skeletal anomalies.⁵

This case is unusual not only because other features of the arhinencephaly spectrum that are often associated with olfactory aplasia were absent, but particularly because this patient was of superior intelligence.

KRISTY E DUNDAS
ROSS Mc D ANDERSON
Department of Anatomical Pathology, Alfred
Hospital, Commercial Road, Prahran, 3181,
Australia

- 1 Louis DN, Arriagada PV, Hyman BT, Hedley-White ET. Olfactory dysgenesis or hypoplasia: A variant in the arhinencephaly spectrum? *Neurology* 1992;42:179-82.
- 2 Kobori JA, Herrick MK and Urich H. Arhinencephaly. The spectrum of associated malformations. *Brain* 1987;110:237-60.
- 3 Adams JH, Duchon LW, eds. *Greenfield's neuropathology*, 5th edn. London: Edward Arnold, 1992:555-9.
- 4 Davis RL and Robertson DM, eds. *Textbook of neuropathology*, 2nd edn. Baltimore, MD: Williams and Wilkins, 1991:225-33.
- 5 Lieblisch JM, Rogol AD, White BJ, Rosen SW. Syndrome of anosmia with hypogonadism (Kallmann's Syndrome), *Am J Med* 1982;73:506-19.

Amoebic brain abscess: a rare but serious complication of *Entamoeba histolytica* infection

Since its first description in 1849,¹ there have been several reports of cerebral amoebiasis.²⁻⁵ To our knowledge only nine adequately documented cases of cure have been described.³⁻⁵ We present a case of cerebral amoebiasis with no evidence of disease elsewhere who made a complete recovery after surgery and treatment with intravenous metronidazole.

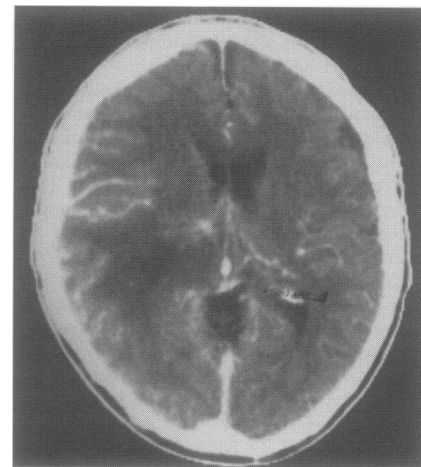


Figure 1 CT scan of the brain with contrast showing an ill defined mass in the right parietal region with ventricular compression.

A 57-year-old man presented to the emergency room with a three day history of progressive weakness of the left half of the body associated with confusion, headache, and vomiting. He had had a sore throat and fever for one week before admission. On admission, he was febrile (39°C) and disorientated. He had a left hemiplegia with reduced sensation, increased tone, and an extensor plantar response on the left side. The rest of the examination was unremarkable. His white cell count was 13.9 with 82% neutrophils and his erythrocyte sedimentation rate was 77. Tests of liver function, blood urea, and creatinine were normal. He was negative for HIV by enzyme linked immunosorbent assay (ELISA) and serum protein immunoelectrophoresis was within normal limits. A chest radiograph showed no abnormality. CT of the head showed a large mixed density lesion in the right posterior parietal region. There was compression of the posterior half of the right lateral ventricle (fig 1). A burr hole biopsy revealed necrotic material, and a provisional report suggested a fungal infection. Treatment with amphotericin B was started.

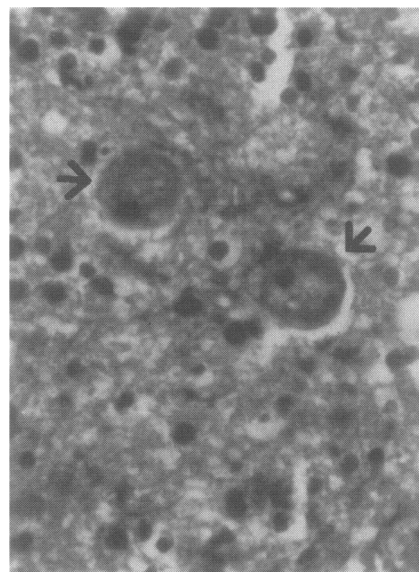


Figure 2 Cerebral cortical tissue showing trophozoites of *Entamoeba histolytica* (periodic acid-Schiff stain, originally $\times 400$).

As the patient continued to deteriorate an emergency craniotomy was made the same evening. The mass consisting of dark brown necrotic material was removed by suction. Widespread cortical thrombophlebitis was noted. Examination of the material removed confirmed the presence of amoebae (fig 2). Treatment with metronidazole (500 mg 6 hours intravenously) was started and continued for three weeks. Postoperatively the patient was ventilated for 48 hours. Thereafter his condition began to improve. Subsequent bacterial and fungal cultures of the biopsy material were negative, whereas ELISA and indirect haemagglutination assay (IHA) titres were 1:3200 and 1:512 respectively.

Investigations to find a primary focus included several stool examinations, which were negative and an abdominal ultrasound, which was normal. Abdominal CT disclosed an irregular, lobulated centrally necrotic mass arising from the upper pole of the right kidney. A needle biopsy of the lesion under ultrasonic guidance was inconclusive. The patient gradually made a complete neurological recovery and was discharged home.

Subsequently the patient underwent a radical right nephrectomy and para-aortic lymphadenectomy for a well differentiated multicystic renal cell carcinoma without extension into perirenal or perineural vascular tissue, or the lymph nodes. There was no evidence of hepatic or renal amoebiasis.

The classic description of the clinical presentation was given by Armitage.⁶ The patient is usually a male in the prime of life who has had dysentery, acute or chronic, with suppurative hepatitis. After liver abscess drainage and while apparently improving, cerebral symptoms and signs develop. The average duration from the first neurological symptom to death is 10 to 15 days. Orbison *et al.*,⁷ in an extensive review

of publications, were able to collect 83 cases in 1961 and added five of their own. The average age of their patients was 32 years. Headache or alteration of mental state were the most common initial presentations. Symptoms and signs of meningitis occurred in 45% of the patients. Abnormalities in the CSF were noted in eight of 10 patients tested. Cranial nerve involvement was frequent. Fever and neutrophilia was noted in most cases; 40% of the patients had no history of amoebic dysentery.

Diagnosis is now facilitated by amoebic serology⁸ and CT scanning⁵ as well as histopathology. The most sensitive test available is IHA; others are latex agglutination, counterimmunoelectrophoresis, indirect immunofluorescence, radioimmunoassay, and ELISA. An IHA titre of 1:128 or an ELISA ranging from 1:1000 to 1:50 000 is suggestive of extra-intestinal amoebiasis. Negative IHA, ELISA, and indirect immunofluorescence make the diagnosis of invasive amoebiasis unlikely.^{8,9} We have found only three previous reports of CT.^{3,5} Becker *et al.*³ described a poorly defined lesion without any zone of reactivity as in our patient. Schmutzhard *et al.* and Tikly *et al.* described small or large ring-like enhancing lesions involving predominantly the white matter with surrounding oedema and midline shift.^{4,5}

Direct observation by light microscopy with an unstained wet preparation will show motile amoebae. Tissue is best obtained from the periphery of the lesion. Permanent Wright stain, Feulgen reaction, and immunofluorescent labelled antibodies directed against the amoebae in tissue may be used or amoebae isolated and cultured, the last requiring considerable experience.

Surgical decompression with removal of necrotic and infected material may be required for the control of intracranial

pressure.^{2,7} Intravenous metronidazole is essential and achieves adequate CNS penetration when the history of illness is relatively short. This report describes only the 10th case documented to have been cured of cerebral amoebiasis with the use of a combined surgical and medical approach.

AHMED ALI SHAH

Department of Neurosurgery

HIZBULLAH SHAIKH

Department of Pathology

MICHELE KARIM

Department of Medicine

The Aga Khan University Medical Centre,
Stadium Road, PO Box 3500, Karachi 74800,
Pakistan

- 1 Morehead C. Notes on the pathology and treatment of diseases of the brain, as observed in the European General Hospital at Bombay. *Transactions of the Medical and Physical Society of Bombay* 1849;9:112-5.
- 2 Lombardo L, Alonso P, Arroyo LS, Brandt H, Mateos JH. Cerebral amoebiasis. Report of 17 cases. *J Neurosurg* 1964;21:704-9.
- 3 Becker GL, Kneep S, Lance KP, Kaufman L. Amoebic abscess of the brain. *Neurosurgery* 1980;6:192-4.
- 4 Schmutzhard E, Mayr U, Rumpl E, Prugger M, Pohl P. Secondary cerebral amoebiasis due to infection with *Entamoeba histolytica*. *Eur Neurol* 1986;25:161-5.
- 5 Tikly M, Denath FM, Hodgkinson HJ, Saffer D. Computed tomographic findings in amoebic brain abscess. *S Afr Med J* 1988;73:258-9.
- 6 Armitage FL. Amoebic abscess of the brain: with notes on a case following amoebic abscess of the liver. *J Trop Med Hyg* 1919;22:69-76.
- 7 Orbison JA, Reeves N, Leedham CL, Blumberg JM. Amoebic brain abscess. *Medicine (Baltimore)* 1951;30:247-82.
- 8 Patterson M, Healy GR, Shabot JM. Serologic testing for amoebiasis. *Gastroenterology* 1980;78:136-41.
- 9 Garcia LS, Bruckner DA, Brewer TG, Shimizu A. Comparison of indirect fluorescent antibody amoebic serology with counter immunoelectrophoresis and indirect hemagglutination amoebic serologies. *J Clin Microbiol* 1982;15:603-5.